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Abstract

Purpose: Provide baseline and preliminary follow-up results in a 5-year longitudinal study of Blau syndrome

Design: longitudinal multicenter prospective observational study

Methods: Baseline data from 50 patients from 25 centers worldwide, and follow-up data for patients followed 1, 2 or 3 years at the end of study enrolment. Ophthalmic data were collected at baseline and yearly visits by means of a standardized collection form.

Results: Median age at onset of eye disease was 60 months and duration of eye disease at baseline 145 months. At baseline 38 patients (78%) had uveitis, bilateral in 37 (97%). Eight patients (21%) had moderate to severe visual impairment. Panuveitis was found in 38 eyes (51%), with characteristic multifocal choroidal infiltrates in 29 eyes (39%). Optic disc pallor in 9 eyes (12%) and peripapillary nodules in 9 eyes (12%) were the commonest signs of optic nerve involvement. Active anterior chamber inflammation was noted in 30 eyes (40%) at baseline and in 16 (34%), 17 (57%) and 11 (61%) of eyes at 1, 2 and 3 year respectively. Panuveitis was associated with longer disease duration. At baseline, 56 eyes (75%) were on topical corticosteroids. Twenty-six patients (68%) received a combination of systemic corticosteroids and immunomodulatory therapy.

Conclusions: Blau uveitis is characterized by progressive panuveitis with multifocal choroiditis, resulting in severe ocular morbidity in spite of continuous systemic and local immunomodulatory therapy. The frequency and severity of Blau uveitis highlight the need for close ophthalmologic surveillance as well as a search for more effective therapies.